

VESICO-APPENDICEAL FISTULA IN A MUCINOUS ADENOCARCINOMA OF THE APPENDIX

Fístula vesíco-apendicular em adenocarcinoma mucinoso do apêndice

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ABSTRACT - Background - A rare case of vesicoappendiceal fistula secondary to mucinous adenocarcinoma of the appendix is presented. **Case report** - A 62-year-old man with a one year history of recurrent urinary tract infections. After two months he developed pneumaturia and fecaluria. An abdominal and pelvic computed tomography demonstrated a trans-mural mass in the posterior wall of the bladder with a vesicoenteric fistula leading to the terminal ileum. Laparotomy revealed a tumor arising from the appendix contiguous with the bladder posterior wall. The bladder was opened and a large fistula and tumor on the posterior bladder wall near the trigone was identified. Frozen pathological analysis showed a mucinous adenocarcinoma. En-bloc right hemicolectomy and partial cystectomy, preserving bladder trigone was performed. After manipulating the tumor, grossly leakage of mucinous materials occurred into the pelvic cavity. A peritoneal washing with a mytomicin solution at 42° C was then performed, to prevent peritoneal seeding. The patient had a prolonged postoperative ileus and was discharged at the 15th day. Five months after the procedure the patient was receiving chemotherapy with 5-fluoracil and leucovorin and there was no signs of recurrent disease. **Conclusion** - The presentation with vesico-appendiceal fistula is extremely rare with only a few cases reported in the literature. Knowledge of different types of neoplasm and appropriate treatment allows the surgeon to provide patients optimal care referring to specialized centers whenever appropriate.

HEADINGS - Appendiceal cancer. Urinary bladder fistula. Adjuvant chemotherapy. Therapeutic hyperthermia.

INTRODUCTION

It's presented a rare case of vesico appendiceal fistula secondary to mucinous adenocarcinoma of the appendix. Patients with adenocarcinoma of the appendix are usually diagnosed with appendicitis, a right lower quadrant abscess, or a tumour mass and many are encountered as incidental findings at laparoscopy or laparotomy^{5,12}. These tumors request different treatments depending on its histology, size, localization in the appendix, grade of invasion, perforation and metastasis. Evolution on adjuvant therapies gives patients and physicians good perspectives in prognosis.

CASE REPORT

A 62-year-old man presented with a 1-year history of dysuria and polaciuria, previously treated with oral antibiotics for recurrent urinary tract infections. He

sought for medical advice and had been submitted to an abdominal ultrasonography, urinary tract ultrasonography and excretory urography which were normal. After two months developed pneumaturia and fecaluria. At physical examination an abdominal mass was palpable at lower right abdominal quadrant. Rectal examination and prostate assessment were normal.

An abdominal and pelvic computed tomography was performed and demonstrated a trans-mural mass in the posterior wall of the bladder with a vesicoenteric fistula leading to the terminal ileum (Figure 1). The CEA (carcinoembryonic antigen) level was 42.

Laparotomy revealed a tumor arising from the appendix contiguous to the bladder posterior wall (Figure 2). The tumor had an intimos relation with right ureter. After opening the bladder a large fistula on the posterior wall near the trigone was identified. The tumor was mucin secreting and frozen pathological analysis showed an appendiceal mucinous adenocarcinoma. It was performed an en-bloc right hemicolectomy and partial cystectomy, preserving bladder trigone and bilateral double loop catheters were passed for adequate urinary drainage. During the procedure it was not observed tumoral peritoneal implants, but after manipulating the tumor, grossly leakage of mucinous materials occurred into the pel-

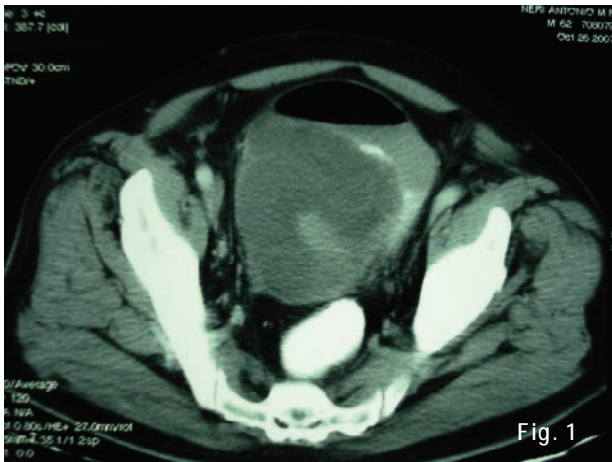


Fig. 1

FIGURE 1 – CT showing a mass contiguous to the urinary bladder and fistula demonstrated by air inside the bladder

vic cavity. Then it was performed a peritoneal washing with a mytomicin solution at 42° C, to prevent peritoneal seeding. Pathological examination demonstrated an exophytic, gelatinous mass with 17 × 12 cm in diameter and vesicoenteric fistula formation (Figure 3). It was classified as a well differentiated mucinous adenocarcinoma of the appendix (Figure 4). Surgical margins were tumor-free and angiolymphatic invasion was not observed. Fifteen dissected lymph nodes were not involved by the tumor. Final pathological diagnosis was a pT4 pN0 pM0 appendiceal mucinous adenocarcinoma.

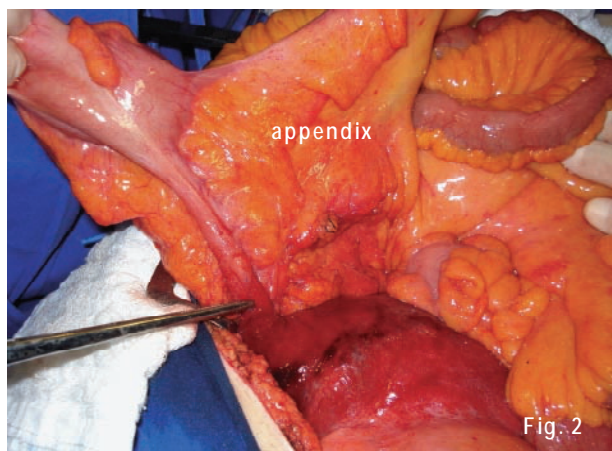


Fig. 2

FIGURE 2 – Intra-operative showing the mass arising from the appendix, invading the urinary bladder

The patient had a prolonged postoperative ileus and received periferical parenteral nutrition from de 5° to the 11° day when adequate oral intake was achieved. At 15th day he was discharged. After two months, it was removed double loop catheters and cystoscopy did not reveal any residual lesion. At five months after the procedure, he was recieving chemotherapy with 5-fluoracil and leucovorin, with no image signs of peritoneal and the CEA remains in normal levels.

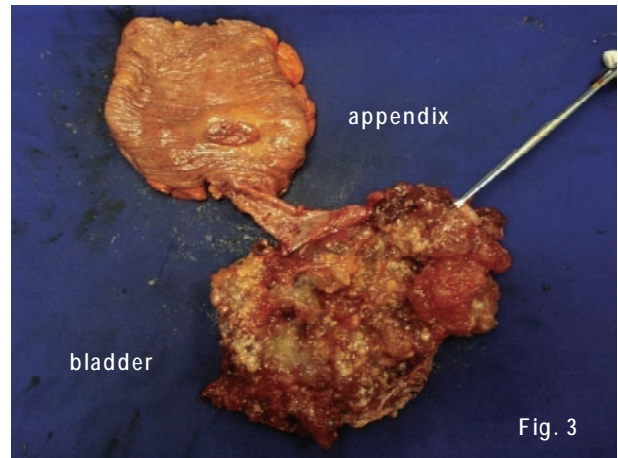


Fig. 3

FIGURE 3 – Surgical specimen after resection

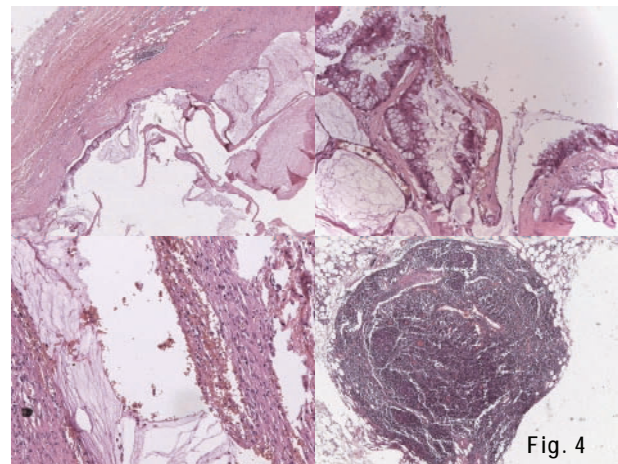


FIGURE 4 – Microscopy showing mucinous adenocarcinoma and bladder invasion

DISCUSSION

Vesicoenteric fistula is usually a complication of an inflammatory or neoplastic process. Common causes include diverticulitis (65-75%), malignant disease (10-15%) and Crohn's disease (5-6%). Presenting symptoms are pneumaturia (52-77%), fecaluria (36-51%) and urinary tract infection symptoms (44-45%)¹¹. Investigation modalities are usually poor, with abdominal CT being the most useful imaging modality. Although cystoscopy can be performed, it is diagnostic in only 30-40% of cases⁸.

Appendiceal carcinomas are rare with an incidence of 0,12 cases per 1,000,000 people per year. It is estimated that appendiceal cancer is found in 1 % of all appendectomy specimens⁶.

Patients with adenocarcinoma of the appendix are usually diagnosed with appendicitis, a right lower quadrant abscess, or a tumour mass^{5,12}. Rarely the tumor can invade the abdominal wall with an enterocutaneous fistula or the urinary bladder with an enterovesical fistula, as shown in this case. International literature report only nine cases of

vesico-appendiceal fistulas derived from mucinous appendiceal adenocarcinomas until present data^{1,2,3,4,8}. This type of tumor usually perforates before diagnosis and may spread to the peritoneal cavity, producing mucinous intraperitoneal ascites, resulting in the pseudomixoma peritonei⁷.

The initial assessment of the incidental appendiceal tumor includes assessment of tumor size, involvement of the base of the appendix or the mesoappendix and its perforation⁷.

If the tumour is confined to the appendix, smaller than 2 cm, without evidence of mesoappendiceal or basal involvement, appendicectomy is the appropriate treatment. An emergency cryostat sectioning should be done and if an invasive non-mucinous adenocarcinoma of the appendix is shown a right hemicolectomy can double the survival achieved with routine appendicectomy¹. In tumours larger than 2 cm or with invasion of the base or mesoappendix, a right hemicolectomy should be performed⁷.

If the appendix has ruptured just before removal or during the operation, it is important to remove all free mucin and perform meticulous peritoneal toilet. After resection, and with the abdomen opened, the peritoneal cavity could be washed with warm (41-5° C) mitomycin solution to avoid peritoneal seeding. Such patients and those who present with a perforated mucinous neoplasm, without evidence of any extra-appendiceal spread, are at risk of developing pseudomixoma peritonei and should be followed up carefully. CT of the abdomen and pelvis and tumour markers (CEA, CA 125 and CA 19.9), provide baseline measure-

ments⁷. At the present case the leakage of peritoneal cavity led to perform the warm mitomycin solution wash. Follow up doesn't show any sign of recurrence, and the patient is been treated with systemic chemotherapy.

Perforated mucinous neoplasm of the appendix with pseudomixoma peritonei syndrome is best treated by complete removal of tumor (peritonectomy) and hiperthermic intraperitoneal chemotherapy [mitomycin solution (41-5° C) during surgery and 5-fluoroucil for four to five days after surgery]⁹.

Right hemicolectomy, without intraperitoneal chemotherapy or complete removal of the tumor, at an initial procedure for a perforated mucinous adenocarcinoma with peritoneal involvement, results in no survival benefit. In this cases the peritoneal cavity should be thoroughly washed out, and the patient should be referred to an appropriate specialized center for definitive treatment^{7,9,12}.

CONCLUSION

Appendiceal neoplasms are uncommon and consists in an heterogeneous group of pathologies. Many present as appendicitis, but some are encountered as incidental findings at laparoscopy or laparotomy. The presentation with vesico-appendiceal fistula is extremely rare with only a few cases reported in the literature. Knowledge of the different types of neoplasm and appropriate treatment allows the surgeon to provide patients optimal care.

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RESUMO - Introdução - Apresenta-se raro caso de fistula vésico-apendicular secundária a adenocarcinoma mucinoso do apêndice. **Relato do caso** - Paciente masculino de 62 anos com história de um ano de infecções urinárias de repetição. Após dois meses desenvolveu pneumatúria e fecalúria, sendo indicada tomografia computadorizada de abdômen que mostrou massa trans-mural na parede da bexiga, com fistula vésico-entérica para região do íleo terminal. A laparotomia revelou tumoração surgindo do apêndice cecal, em continuidade com a parede posterior da bexiga. A mesma foi aberta, sendo identificada grande fistula e material tumoral até próximo ao trígono vesical. A biópsia de congelação identificou adenocarcinoma mucinoso. Realizada hemicolectomia associada à cistectomia parcial em bloco, com preservação do trígono vesical. Houve extravasamento de mucina para a cavidade pélvica pela manipulação. Optado por lavagem peritoneal com solução de mitomicina a 42° C para prevenir disseminação peritoneal. O paciente evoluiu com íleo prolongado pós-operatório, tendo alta hospitalar no 15° dia. Cinco meses após o procedimento encontrava-se em quimioterapia com 5-fluoracil e leucovorin, sem sinais de doença recidivante. **Conclusão** - A presença de fistula vésico-apendiceal é extremamente rara, tendo poucos casos relatados na literatura. O conhecimento dos diferentes tipos de neoplasias e seus tratamentos adequados permite ao cirurgião oferecer melhor cuidado ao paciente.

DESCRIPTORIOS - Câncer do apêndice. Fístula urinária. Quimioterapia adjuvante. Hipertermia terapêutica.

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